Desmoplastic fibroblastoma (collagenous fibroma) is a fibrous, soft-tissue tumor first described by Evans (1) in 1995. This tumor is clinically and morphologically distinct and was deemed completely benign in a few reported series (1–3). There have also been published case reports on the MR features of desmoplastic fibroblastoma (4–8). We herein present a case of desmoplastic fibroblastoma with special focus on the MR findings, because they are distinct from that previously published.

Case Report

A 33-year-old female presented with an eight-month history of a palpable mass in her right arm without associated neurologic symptoms. Physical examination revealed a hard, well-circumscribed and movable mass measuring 4×4×2 cm. Plain radiography of the mass demonstrated no calcification (not shown).

Gray-scale imaging on ultrasonography showed a hypoechoic mass lesion with internal vascularity. Magnetic resonance (MR) images showed areas of heterogeneously with high and low signal intensity on T2-weighted images, and intense enhancement on post-contrast T1-weighted images. Histopathologic examination of the resected specimen confirmed desmoplastic fibroblastoma.

Index words: Soft Tissue Neoplasms
Fibroblastoma, Desmoplastic
Magnetic Resonance Imaging
Fig. 1. A 33-year-old female with a $4 \times 4 \times 2$ cm mass in the anterior aspect of the right arm.
A. Color Doppler imaging shows a well-defined, hypoechoic mass with vascular signals in the peripheral and central portions of the mass.
B. Axial FSE T1-weighted MR image (TR/TE = 500/20) shows slightly high signal intensity of the mass compared to that of the adjacent muscles [arrows]. The mass is located in the subcutaneous layer and is attached to the right biceps brachii and brachialis muscles.
C. Coronal FSE T2-weighted MR image (TR/TE = 2141/100) shows slightly low signal intensity with high signal intensity in the upper area within the mass [arrows].
D. Axial FSE T1-weighted MR image (TR/TE = 602/20) obtained following contrast injection, shows marked enhancement of the mass [arrows]. The mass shows mild indentation to the adjacent muscles with no evidence of invasion.
E. H & E staining ($\times 100$) shows spindle to stellate fibroblastic cells in a hypovascular collagenous matrix.
sional biopsy. There was no evidence of invasion to the muscles of her right arm, and there were no tumor cells in the resected margin of the frozen section.

Macroscopically, the tumor was firm without cystic or hemorrhagic changes. Microscopically, the tumor was composed of spindle to stellate fibroblastic cells in a hypovascular collagenous matrix (Fig. 1E). The immunohistochemical study was negative on S-100 protein. The final diagnosis was desmoplastic fibroblastoma.

The patient experienced no signs of recurrence and remained healthy 16 months after surgery.

Discussion

Desmoplastic fibroblastoma (collagenous fibroma) is a benign fibrous tumor most often seen in patients during the fifth and sixth decades of life, with a male to female ratio of 5:1. Most tumors occur in the extremities, shoulder girdle, posterior neck, upper back and abdominal wall. The size of reported desmoplastic fibroblastomas ranges from 1 to 20 cm in maximal diameter. The lesion is typically a well-circumscribed, firm mass that involves the subcutaneous or deep soft tissue [1-3].

Histologically, the tumor cells are relatively bland stellate- and spindle-shaped fibroblastic cells, separated by a densely fibrous to fibromyxoid matrix. The cellularity ranges from low to very low. The mitotic figures are very low or completely absent. Tumor necrosis is not seen [1-3].

There have been a few case reports of desmoplastic fibroblastoma with MR features [4-8]. Three cases of desmoplastic fibroblastoma in the upper extremity, and one case in a hip joint showing diffusely low signal intensity of the mass on T2-weighted images [4, 5, 7] have been reported. One reported case of desmoplastic fibroblastoma in the peritoneal cavity showed marked heterogeneous signal intensity on T2-weighted and post-contrast T1-weighted images [6]. These findings are slightly different from those seen in our case, which had heterogeneously high and low signal intensity on T2-weighted images, and well-enhanced post-contrast T1-weighted images. According to a recent report of desmoplastic fibroblastoma with erosion of the right L5 pedicle, MR images showed heterogeneous intermediate signal intensity on T2-weighted image with scattered areas of low signal intensity, and minimally heterogeneous enhancement [8]. However, our case showed higher heterogeneous signal intensity on T2-weighted images and more prominent enhancement.

Most soft-tissue masses have high signal intensity on T2-weighted images. In the absence of calcification, abundant collagen and marked hypocellularity in a soft-tissue tumor resulted in reduced signal on T2-weighted pulse sequence [9]. The area showing high signal intensity on T2-weighted images corresponded to the hypercellular area within the lesion, consisted of a tumor with loose collagen fibers [6]. The high signal intensity on T2-weighted images seen in our case is probably due to high cellularity and small amount of loose collagen fibers in the mass.

The radiologic differential diagnosis included other soft-tissue neoplasms, such as desmoid tumor, neurogenic tumor and soft-tissue sarcoma. Among these differential diagnosis, desmoid tumor is the most important consideration for clinicians. Histologically, desmoid tumor has a greater infiltrative potential and is usually more cellular than desmoplastic fibroblastoma. On MR imaging, desmoid tumor may also have larger areas showing very high signal intensity on T2-weighted images, thus indicating higher cellularity than desmoplastic fibroblastoma [10]. If high signal intensity on T2-weighted images is prominent on MR imaging, as was in our case, differentiating desmoplastic fibroblastoma from desmoid tumor becomes very difficult.

Although desmoplastic fibroblastoma is usually revealed by low signal intensity on T2-weighted MR images, it may appear as a well-defined soft-tissue mass with heterogeneously high and low signal intensity on T2-weighted images, and as intense enhancement on MR imaging.

Acknowledgments

We express our sincere gratitude to Bonnie Hami of the Department of Radiology, University Hospitals of Cleveland, for her editorial assistance in the preparation of this manuscript.

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대한영상의학회지 2010;63:267-270

결합조직형성섬유아세포종의 자기공명영상 소견: 증례 보고1

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이원찬∙강병성∙서재희2∙황재철∙원영철∙신상훈∙권운정∙최성훈

결합조직형성섬유아세포종은, 보통 다양한 부위의 피하조직 또는 근육조직에서 생기는 드문 섬유성 연조직 종양이다. 저자들은 우측 상완부의 만져지는 종괴를 주소로 내원한 33세 여자 환자를 검사하였다. 초음파 검사에서 내부의 혈류를 보이는 저에코의 종양이 있었다. 자기공명영상 검사에서는 T2 강조영상에서 불균질한 고신호와 저신호강도를 보였으며 조영증강 T1 강조영상에서 강한 조영증강을 보였다. 증례는 조직병리학 검사에서 결합조직형성섬유아세포종으로 확진되었으며 저자들은 결합조직형성섬유아세포종의 자기공명영상 소견을 보고하고자 한다.